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Ramsay Hunt Syndrome: a case report

Síndrome de Ramsay Hunt: relato de caso

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ABSTRACT

Ramsay Hunt syndrome is a disorder characterized by herpetic eruptions on the auricle and facial paralysis, being the infection in the geniculate ganglion attributed to varicella zoster virus. The purpose of the article is to describe the clinical characteristics of diagnosis and treatment of a patient with Ramsay Hunt syndrome. A male patient, 72 years old, sought treatment for facial and neck pain, grade 10, on the visual analog scale (VAS = 10). The patient claimed that he had the pain for 15 days and described it as a short term electric shock, involving the auriculotemporal branch of the trigeminal nerve and the great auricular nerve of the superficial cervical plexus on the right side. Besides the pain, hemifacial paralysis on the same side of the face was also observed. Because of the location, quality and duration of the pain, associated with the clinical manifestations, acyclovir therapy and physiotherapy were the treatments proposed. In the third follow-up visit, the patient reported no facial and neck pain, EAV = zero, although there was still a slight asymmetry of the buccal branch of the facial nerve on the right side. The Ramsay Hunt syndrome is a disease that can, in some cases, lead to hemifacial paralysis, causing the patient's individual retraction of social life. Both early diagnosis and treatment seem to be directly related to therapeutic success.

KEYWORDS: Facial nerve; Facial paralysis; Herpes zoster oticus.

RESUMO

A síndrome de Ramsay Hunt (SRH) é uma doença caracterizada por erupções herpéticas auriculares e paralisia facial, atribuída a infecção pelo varicela-zoster vírus (VZV) no gânglio geniculado. O objetivo do artigo é descrever as características clínicas, diagnóstico e tratamento de um paciente com síndrome de Ramsay Hunt. Um paciente do sexo masculino, 72 anos, procurou tratamento para dor cérvico-facial, de grau 10, pela escala analógica visual (EAV = 10), que persistia durante 15 dias. Relatou uma dor do tipo choque elétrico, de curta duração, envolvendo o nervo auriculotemporal do trigêmeo e o nervo auricular maior do plexo cervical superficial no lado direito. Associada com a dor estava presente uma paralisia hemi facial do mesmo lado da face. Devido a localização da dor, qualidade, duração e manifestações clínicas, optou-se por um tratamento a base de aciclovir e fisioterapia. Na terceira consulta de revisão, o paciente não apresentava mais dor cérvico-facial, EAV = zero, mantendo-se ainda uma leve assimetria do ramo bucal do nervo facial no lado direito. A síndrome de Ramsay Hunt é uma patologia que pode, em alguns casos, levar a paralisia hemifacial e desencadear por parte do portador da mesma uma retração do convívio social. O diagnóstico e tratamento precoces parecem estar relacionados diretamente ao sucesso terapêutico

PALAVRAS-CHAVE: Nervo facial; Paralisia facial; Herpes zoster da orelha externa.

INTRODUCTION

The Ramsay Hunt syndrome (RHS) is a herpes infection in the geniculate ganglion of the seventh cranial nerve, caused by reactivation of the latent form of varicella-zoster virus[1]. Patients usually have facial palsy, ipsilateral otalgia and hearing loss or tinnitus accompanied by vesicular lesions [2]. Cranial nerves VIII, IX, V, VI and X (in this order of frequency) may also be involved [3]. The syndrome may manifest itself at any time after the patient is infected with the virus, but it is more common in adults and in individuals with impaired cell-mediated immunity [4]. It is considered potentially serious, because complete recovery cannot take place once nerve degeneration has occurred and it represents about 12% of all facial nerve palsies [5].

The clinical diagnosis of RHS is made when there are clinical findings of unilateral

rash or blisters on the facial region associated with unilateral facial paralysis and severe facial pain. The infection can be diagnosed by analyzing the vesicular fluid by Tzanck smear, viral culture, PCR (Polymerase chain reaction) or fluorescence microscopy. Serological methods for diagnosis are based on an increased IgG index or the finding of specific IgM antibodies [3] The purpose of the article is to describes the clinical features, diagnosis and treatment of a patient with Ramsay Hunt syndrome, which was referred to a maxillofacial surgery department.

CASE DESCRIPTION

Male patient, 72 years old, sought treatment for facial and neck pain, Grade 10 on the visual analog scale (VAS = 10), which started and persisted for about 15 days. The anamnesis revealed that the patient had vesicular and bullous lesions in the external ear and external auditory canal; however they had already healed when the clinical exam was performed by the authors. He also reported felling pain, describing it as a short-term electric shock involving the auriculotemporal branch of the trigeminal nerve and the great auricular nerve of the superficial cervical plexus on the right side of the face.The patient reported no prior knowledge of infection with varicella zoster virus. He was in good health and reported being under treatment for hypertension. Besides the pain, hemifacial paralysis on the same side of the face was also observed. (Figure 1).

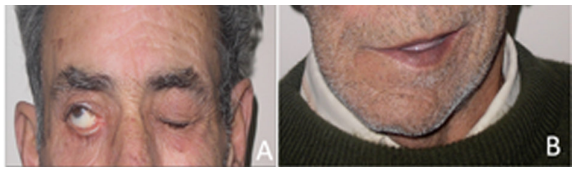

Figure 1 A- Loss of motor function of temporal and zygomatic branches of the facial nerve on the right. B - Loss of motor function of buccal branch of facial nerve on the right

The patient had already been submitted to a tomography of the skull base, audiometric tests, impedance, and other tests that did not show any abnormality..

The clinical diagnosis was established as Ramsay Hunt Syndrome based on the location, quality and duration of the pain associated with the clinical manifestations..

The treatment proposed consisted of Acyclovir 800 mg five times a day for 10 days. Physiotherapy treatment was also recommended for the same period of the drug therapy. In the third follow-up visit, the patient reported no facial and neck pain, EAV = zero, although there was still a slight asymmetry of the buccal branch of the facial nerve on the right side of the face (Figure 2).

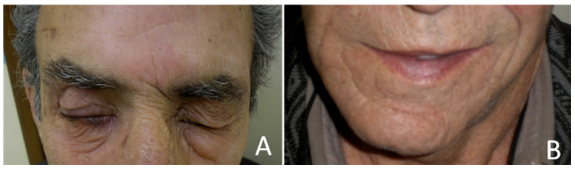

Figure 2- A Return of motor function in the upper third of the face. B - Return incomplete of the motor nerve of the buccal facial nerve on the right.

DISCUSSION

The varicella-zoster virus (VZV) belongs to the human herpes virus group and it is the etiologic agent of varicella and herpes zoster [6, 7]. This virus is a ubiquitous agent of infection and it is known that at least 95% of the adult population is infected [8]. It may be latent in the nerve root ganglion for years in a patient who had previously had chickenpox. However, some factors can reactivate it, especially when an individual's immunity decreases, causing herpes zoster. The infection by the virus of the geniculate ganglion of the facial nerves causes herpes zoster oticus also known as Ramsay Hunt syndrome (RHS), causing ear pain, facial paralysis and homo or bilateral vesicular eruptions in the outer ear and ear canal. The patient described here did not report any hearing impairment, but he suffered of severe pain in the ear. In severe cases of RHS, the involvement of the cochlear nerve leads to hearing loss in 10% of the cases [6].

The clinical diagnosis of RHS is made primarily when erythematous vesicles in one facial dermatome are found in association with unilateral facial paralysis and severe facial pain[1]. In this case the patient reported not being aware of a prior infection with varicella zoster virus. The diagnosis was based on these clinical signs and symptoms. Serologic tests to confirm theinfection by varicella zoster virus are not necessary, even if other cranial nerves are affected [9]. The patient did not present vesicular lesions at the time of the physical examination, but he reported they had appeared accompanied

by facial paralysis and pain, which were still present during the exam.

The traditional treatment for RSH has consisted in the of corticosteroids in high doses, however this approach has been changing with the development of new virostatic agents, especially acyclovir. Acyclovir has a good virostatic action among different types of herpes, interfering with DNA polymerase and inhibiting their replication [5]. Therefore, the best results are obtained when administered at disease's onset [5]. In this case, we used the correct medication in the appropriate dose, but the lack of an earlier diagnosis probably resulted in the only partial recovery of motor function of the facial nerve. The literature reported histological analysis of patients with RHS that showed areas of atrophy and nerve degeneration[10], which may have occurred in our case regarding the buccal branch of facial nerves on the right side of the face.

Sometimes antiviral drugs are associated with corticosteroid anti-inflammatory drugs [2,3], being this therapy recommended to improve the recovery of the facial nerve function in patients with RHS [11], not being reported side effects in relation to this treatment [5]. In this case, acyclovir produced, in a short period of time, the desired effects, being an antiviral administration and no steroids, because the patient was under treatment for hypertension. In patients with diabetes, the treatment with corticosteroids should be avoided and only NSAIDs should be prescribed [6]. Antibiotic ointments and sprays can be used in facial vesicular lesions [12].

The pain in patients with RHS can last for up to 2 months [13]. In this case, the pain has persisted for 15 days. We tried to measure it using a visual analog scale (VAS) from the beginning to the end of the treatment, and it allowed us to verify the complete remission of the painful symptoms after 10 days of treatment.

CONCLUSION

The Ramsay Hunt syndrome is one of the cause of not traumatic acute peripheral facial palsy, so the professionals must be vigilant for signs and symptoms. Early treatment of this syndrome provides a complete clinical recovery, avoiding sequelae of irreversible character, due to nerve degeneration.

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